CLINICAL CARE MANAGEMENT GUIDELINE

ANNUAL NEUROLOGICAL EXAMINATION FOR SUPERFICIAL SIDEROSIS

Version 1.1
08.05.23
Reviewed: Michael Levy MD, PhD

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INTRODUCTION

Superficial Siderosis is a rare neurodegenerative disorder characterized by the deposition of hemosiderin (a breakdown product of hemoglobin) on the surface of the brain and spinal cord. This complex condition can present with a variety of symptoms, affecting both the central and peripheral nervous systems.

Given the multifaceted nature of Superficial Siderosis, a thorough annual neurological examination is critical for ongoing management and monitoring of the patient. The following guidelines are designed to provide an organized and detailed assessment.

PREPARATION FOR THE EXAM

REVIEW PREVIOUS IMAGING STUDIES

Ensure that MRI sequencing has been completed prior to the exam, so there is adequate time to review and familiarize yourself with the patient's situation.

PATIENT COMMUNICATION

Explain the examination process to the patient, outlining the importance of their feedback and cooperation.

INTERDISCIPLINARY APPROACH

Coordination with other medical professionals such as audiologists, urologists, gastroenterologists, etc., can provide a more comprehensive understanding of the patient's condition.

MRI IMAGING PROTOCOLS FOR SUPERFICIAL SIDEROSIS EVALUATION

WHOLE BRAIN MRI WITH SWI (SUSCEPTIBILITY WEIGHTED IMAGING) WITHOUT CONTRAST

Reason for Exam: Evaluate for evidence of superficial siderosis.
Special Instructions: Please include axial, coronal, and sagittal views. Pay special attention to the cortical sulci and cerebellum. Compare to previous studies for change in hemosiderin deposition.

WHOLE SPINE T2-WEIGHTED IMAGING

Reason for Exam: Evaluate for evidence of superficial siderosis.
Special Instructions: Please include axial and sagittal views of the cervical, thoracic, and lumbar spine. Compare to previous studies for change in hemosiderin deposition.

WHOLE BRAIN MRI WITH VOLUMETRIC SEQUENCES

Reason for Exam: Evaluate for evidence of cerebellar atrophy or volume loss.
Special Instructions: Please include 3D T1-weighted sequences for volumetric analysis. Axial, coronal, and sagittal views should be included. Focus on the cerebellum and brainstem. No contrast is to be used for this study.

EXAMINATION GUIDELINES

MENTAL STATUS: 5 MINUTES

1. Allow the patient to give a 5-minute monologue (unprompted) about the previous year's medical history. This will help assess their mental state and cognition. If there are obvious cognitive dysfunction, then:
    a. Order Montreal Cognitive Assessment (MoCA): 20 Minutes in office
    b. If ≥ 3pt decline of total MoCA score from previous exam refer for neurophysiological testing.

CRANIAL NERVES: 5-10 MINUTES

1. Assess visual fields by confrontation.
2. Check pupils and eye movements.
3. Observe the patient for asymmetries during natural speech and symmetry of eye blinks.
4. Lower cranial nerves (IX-XII) should only be tested if dysphagia and dysarthria are present.

MOTOR EXAMINATION: 10-15 MINUTES

1. Watch for the presence of adventitial movements such as tics, tremors, and bradykinesia.
2. Perform the pronator drift test to identify potential upper motor neuron dysfunction.
3. Look for external rotation of the leg, which may imply upper motor neuron dysfunction.
4. Assess muscle tone, which is important for noting subtle upper motor neuron dysfunction.
5. Conduct functional strength testing, which is preferred over formal push-pull testing.

SENSORY EXAMINATION: 5-10 MINUTES

1. Focus sensory testing on the patient's known symptoms, as sensory testing is purely subjective.
2. Check for a sensory level on the back if hemosiderin deposition is displayed on spinal cord imaging.
3. Perform the test of touching the nose with eyes closed.
4. Conduct the Romberg test for proprioception (peripheral nerves).

COORDINATION: 5-10 MINUTES

1. Assess ataxia and truncal stability.
2. Observe mental status, adventitial movements, and facial symmetry.

REFLEXES: 3-5 MINUTES

1. Look for asymmetries and sustained clonus, which is a purely objective assessment.
2. Check for the Babinski sign, but do not over-interpret the results.

GAIT: 5-10 MINUTES

1. Assess the patient's base, stride, arm-swing, turns, and symmetry.
2. Evaluate the patient's gait under different conditions: casual, heel, toe, and tandem walking.

PROGRESSION ASSESSMENT

Here are some questions that could be asked during the annual superficial siderosis assessment to track progression changes in the mentioned conditions. The patient's responses should guide further questioning and examination. It's also important to ask about any other symptoms or concerns the patient may have, even if they don't seem directly related to superficial siderosis.

URINARY FUNCTION

Have you noticed any changes in your urinary habits, such as frequency, urgency, or incontinence?

BOWEL FUNCTION

Have you noticed any changes in your bowel movements, such as frequency, consistency, or incontinence?
Have you experienced any abdominal pain, bloating, or discomfort?

HEADACHES

Have you experienced any headaches in the past year?
If so, can you describe the location, intensity, and duration of these headaches?
Have there been any changes in the frequency or severity of your headaches?

GASTROESOPHAGEAL REFLUX DISEASE (GERD)

Have you experienced any heartburn or acid reflux symptoms?
Have you noticed any changes in these symptoms, such as increased frequency or severity?

SLEEP APNEA

Have you or anyone else noticed that you stop breathing during your sleep?
Do you snore loudly or wake up gasping for air?

FATIGUE

Have you felt more tired or fatigued than usual?
Has this fatigue interfered with your daily activities or quality of life?
Have you noticed any patterns or triggers for this fatigue?

PERIPHERAL NEUROPATHY

Have you experienced any numbness, tingling, or pain in your hands or feet?
Have you noticed any changes in these symptoms, such as increased frequency or severity?
Have these symptoms affected your ability to perform daily activities?

RADICULAR PAIN

Have you experienced any sharp, shooting pains that travel down your arms or legs?
Have you noticed any changes in these symptoms, such as increased frequency or severity?
Have these symptoms affected your ability to perform daily activities?

POST-EXAMINATION CONSIDERATIONS

DOCUMENTATION

Detailed record-keeping is vital for monitoring changes over time.

PATIENT EDUCATION

Provide the patient with a clear understanding of their progression status, potential therapies, and preventative measures.

FOLLOW-UP AND REFERRALS

Determine the need for further testing or specialist consultations.

COORDINATION WITH OTHER CARE PROVIDERS

If applicable, ensure that all healthcare providers involved in the patient's care are informed of the findings and any changes in management.

EMOTIONAL SUPPORT

Recognize and address any emotional concerns or anxieties the patient may have about their condition and its progression.

CONCLUSION

Superficial Siderosis is a complex and heterogeneous disorder, and the outlined guideline offers an essential framework for conducting a comprehensive annual neurological assessment. By focusing on individualized evaluation and monitoring, it encourages patient-centric care that can adapt to the unique characteristics and challenges of each patient's condition. Utilizing state-of-the-art imaging techniques and a multidisciplinary approach further enhances the accuracy and effectiveness of diagnosis and ongoing management.

The careful consideration of all these factors can result in improved patient outcomes and quality of life, even in the face of this challenging neurological condition.